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## **How I treat acquired Factor VIII inhibitors**

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## **Abstract**

Acquired hemophilia A is a rare bleeding diathesis caused by autoantibodies directed against clotting factor VIII and associated with an increased morbidity and mortality. This autoimmune disorder most commonly occurs in the elderly. Although it may be associated with several underlying pathologies, up to 50% of reported cases remain idiopathic. In contrast with congenital hemophilia, which is commonly characterized by hemarthroses, hemorrhages in patients with acquired hemophilia involve most frequently soft tissues. The two treatment priorities are to arrest the acute bleeding and to eradicate the factor VIII autoantibody. Acute bleeding episodes in patients with low-titer inhibitors can be treated using human factor VIII concentrates, whereas factor VIII bypassing agents, such as activated prothrombin complex concentrates or recombinant activated factor VII, are effective for the treatment of those with high-titer inhibitors. An analysis of the literature shows that the most effective first-line treatment for the eradication of factor VIII autoantibodies is the combination of steroids and cyclophosphamide. However, there is increasing evidence on the effectiveness of other treatment approaches, such as immune tolerance regimens and rituximab. If confirmed by large controlled studies, these innovative therapies might become a valid option for long-term eradication of factor VIII inhibitors.

**Key words:** autoantibodies, factor VIII, hemophilia, inhibitors.